PAPER

A population based study of intracranial arachnoid cysts: clinical and neuroimaging outcomes following surgical cyst decompression in adults

Christian A Helland, Knut Wester

J Neurol Neurosurg Psychiatry 2007;**78**:1129–1135. doi: 10.1136/jnnp.2006.107995

See end of article for authors' affiliations

Correspondence to: Dr Christian A Helland, Department of Neurosurgery, Haukeland University Hospital, N-5021 Bergen, Norway; chhe@helse-bergen.no

Received 30 September 2006
Revised 12 January 2007
Accepted 18 January 2007
Published Online First 13 February 2007

Background: We have gradually adopted a liberal attitude towards surgical decompression of arachnoid cysts. This study describes the results from our institution.
Methods: Long term clinical and neuroimaging results of 156 adult patients (aged ⩾16 years) operated on for arachnoid cysts in our department during the period January 1987 to September 2004 were assessed based on their medical and neuroimaging records, and on a questionnaire.
Results: The clinical and/or neuroimaging results indicated that the cyst was successfully decompressed in all patients. 82% of patients were asymptomatic or had insignificant complaints at follow-up. 12% reported no symptom relief whereas 6% experienced worsening of symptoms. The cyst disappeared after surgery, or was reduced to <50% of the preoperative volume, in 66% of cases. In another 24%, the postoperative volume was also reduced, but was larger than 50% of the original cyst volume. No reduction in fluid volume was observed in 10% of cases. There was no association between volume reduction and clinical improvement. A complication occurred in 26 patients (17%), all with temporal cysts, leading to reoperation in 11 patients (7.1%). In only two patients did the complication cause a permanent slight disability.
Conclusion: Decompression of arachnoid cysts yields a substantial clinical benefit with a low risk of severe complications.

Arachnoid cysts are benign congenital malformations of the arachnoid. They can be located along the craniospinal axis, with a predilection for the temporal fossa.[1] They may present with specific symptoms, such as sensorimotor symptoms corresponding to the location of the cyst, but more often they yield unspecific symptoms (eg, headache or dizziness, or symptoms related to suboptimal cerebral function, such as epilepsy or impaired cognition).[2–10]

The treatment of such cysts, particularly in the majority of the patients with moderate and unspecific symptoms, has been controversial.[11–15] Many authors have expressed a reluctance to operate on these patients unless the symptoms are dramatic. This was also the initial attitude of the senior author. However, based on several observations of our own and those of others, we have gradually changed our view. When we encountered patients that were severely impaired, with symptoms such as headache or dizziness, and who also had radiologically expansive cysts, we found it logical to attempt to alleviate the complaints by surgical cyst decompression. It has been our accumulated experience that surgical decompression yields clinical improvement in most patients with arachnoid cysts. Also, a growing literature indicates that cyst decompression improves the function of neighbouring cerebral tissue, thus supporting the view that patients with unspecific symptoms and ''clinically silent'' cysts may also profit from surgical cyst decompression.[4 6–10 16]

Treating a benign condition that is not life threatening with surgery, with the aim of improving quality of life, can only be justified when a clear clinical benefit, and no severe complications, can be demonstrated. We therefore wished to describe our experience with a relatively liberal indication for surgical decompression of intracranial arachnoid cysts. We have recently published a similar study on our paediatric cyst patients.[17]

MATERIAL AND METHODS

Hospital structure and study population

During the study period, Norway had a three level hierarchical hospital structure, with local community hospitals as the primary referral centre. As a secondary referral centre, most counties had a central county hospital that could refer patients to one of the five university clinics with a neurosurgical department, each serving a health region. Only the university clinics had neurosurgical departments. A health region consists of several counties, with a well defined population and little or no overlap between the health regions. Haukeland University Hospital is the university hospital on the south-western coast of Southern Norway, serving three counties with a total population of 930 000 (2003). During the study period, a few patients were referred to our department from other health regions for surgical cyst decompression, but cyst patients from our region were not referred to other regions.

Patients

A total of 156 consecutive patients (95 males and 61 females) that had been operated on by the senior author (KW) were included in the study. Mean age was 39.0 years (median 36.5; range 16.1–74.5). Mean follow-up duration was 70.5 months (median 67.7, range 4–177). Thirty-four of the patients were treated with a special surgical technique (a cysto-subdural shunt). They are presented in more detail in a separate study.[18]

The majority of patients had a temporal cyst (113, 72.4%) or a frontal cyst (24, 15.4%). Thirteen patients (8.3%) had a posterior fossa cyst while six patients had cysts in other different locations. The main preoperative complaints and cyst locations are summarised in tables 1 and 2.

Abbreviations: COG, clinical outcome group; NOG, neuroimaging outcome group; SDH, subdural haematoma

Table 1 Main preoperative complaint for different cyst locations

	Location of cyst			
Complaint	Temporal (%)	Frontal (%)	Posterior fossa (%)	Other (%)
Headache	88 (77.9)	15 (62.5)	11 (92.3)	2 (40.0)
Epilepsy	14 (12.4)*	8 (33.3)†	1 (7.8)	3 (60.0)
Dizziness/nausea	27 (24.8)	2 (8.3)	8 (61.5)‡	2 (40.0)
Dyscognition	8 (7.1)	4 (16.7)	1 (8.3)	

Some patients had more than one complaint
*Less frequent than for other locations (p=0.029).
†More frequent than for other locations (p=0.005).
‡More frequent than for other locations (p=0.019).

Study design and data extraction

The present study is a questionnaire based retrospective study that includes all adult patients (aged >16 years) that were operated on for an arachnoid cyst in our department during the period January 1987 to September 2004. The patients were identified from the hospital's computer bank of diagnosis and treatment, with registration of every patient in the period with a diagnosis code 348.0 (ICD 9) or G 93.0 (ICD 10), the diagnostic codes assigned to patients with arachnoid cysts in our hospital during the study period. These codes also include other intracranial cysts, but only patients with a verified arachnoid cyst were included. A total of 194 adult patients with arachnoid cysts were seen by the authors during this period; thus 38 patients were not operated on.

A cross check was performed with the handwritten OR protocols from the period, as well as the typed surgical records kept by the senior author (KW), in order to ensure that all adult patients operated on for an arachnoid cyst were included.

All patients selected for surgery underwent a cerebral CT or MRI scan prior to the operation, and a postoperative scan within the first 72 h after surgery. Patients were routinely readmitted 3–6 months after surgery for a postoperative control, including clinical examination and a CT or MRI scan. Patients with any postoperative neuroimaging abnormalities (eg, subdural haematoma or hygroma) were followed further until spontaneous remission or until remission occurred after surgical treatment of the complication.

The following data were extracted from the complete medical records of the operated patients: demographic data, presenting complaint(s), clinical findings, location of the cyst, Galassi type for the temporal cysts,[19] side, postoperative change in cyst size, surgical method and possible clinical or neuroimaging adverse effects of the operation, such as subdural hygromas or haematomas, or new neurological symptoms/deficits.

During the Autumn of 2004, a personal letter explaining the purpose of the investigation was sent to each patient. Patients were asked to complete an enclosed tick box questionnaire, and return it in a preaddressed prestamped envelope. The questionnaire included questions on changes in the preoperative complaint(s) after the operation, visual analogue scales for grading the present level of preoperative complaint(s), level of function in everyday activities, whether the patient had any regrets about undergoing surgery and whether the surgery had produced any new complaints. There was also ample space after each question for the respondent to provide supplementary information.

Table 2 Cyst location

Cyst location	Side	No. of patients (%)
Temporal fossa	Left	77 (49.4)
	Right	33 (21.2)
	Bilateral	3 (1.9)
Frontal	Left	8 (5.1)
	Right	16 (10.3)
Posterior fossa	Left	1 (0.6)
	Right	5 (3.2)
	Midline	7 (4.5)
Other locations		6 (3.8)
Total		156 (100)

Selection criteria for surgery

With two exceptions, all patients that were offered an operation for their arachnoid cyst had one or more complaints that could be related to the cyst. In the two asymptomatic patients, one had a chronic subdural haematoma caused by the cyst and the other had a very large type III cyst with pronounced compression of the underlying brain and midline shift.

Surgery

In the majority of patients (122, 78.2%), a craniotomy was performed under general anaesthesia. During this operation, the parietal cyst membrane covering the inside of the dura was pealed off the dura and then removed from its attachment to the surrounding cortex, for temporal cysts all the way to the tentorial slit. The arachnoid membrane that covered the brain surface (the visceral membrane) was not removed. Any bridging veins running for some distance along the parietal membrane before they drained into the basal sinuses were coagulated and removed in order to prevent postoperative oozing of blood from where the veins drained into the basal sinuses. Large bridging veins near the sphenoid ridge, running for only a very short distance along the membrane before draining and believed to be supported by the surrounding tissue in such a way that they would not move and cause a haemorrhage, were not extirpated. The medial, remaining membrane, covering the basal structures (the tentorial slit, the oculomotor nerve, the carotid artery and the optic nerve) were fenestrated, thus creating communication to the basal cisterns and the posterior fossa. When accessible, the arachnoid covering the Sylvian fissure was also opened, thus creating communication to the subarachnoid space surrounding the carotid and middle cerebral arteries. In 34 patients (21.8%), the cyst was primarily shunted from the cyst to the subdural space with insertion of an internal shunt catheter to the subdural space. This was performed as a minimally invasive procedure through a trephine hole under local anaesthesia. The method has been described in detail elsewhere.[20]

Scoring of clinical results

The clinical results of surgical cyst decompression were categorised into one of four clinical outcome groups (COGs) based on reports from the medical journal and the questionnaire:

- the preoperative complaints had disappeared entirely or were negligible (COG1);
- the preoperative complaints were clearly reduced but still present (COG2);
- the preoperative complaints were unchanged (COG3);
- the patient had more complaints after operation (COG4).

Scoring of neuroimaging results

The neuroimaging results of the decompression, measured as the change in cyst volume on the neuroimaging examination after 3–6 months, were categorised into one of four possible neuroimaging outcome groups (NOGs) (fig 1):

- the cyst had disappeared and was no longer visible (NOG1);
- a fluid volume was still visible where the cyst had been, smaller than 50% of the preoperative cyst volume (NOG2);
- as above, but the residual volume was larger than 50% of the preoperative cyst volume (NOG3);
- no change in cyst volume could be observed (NOG4).

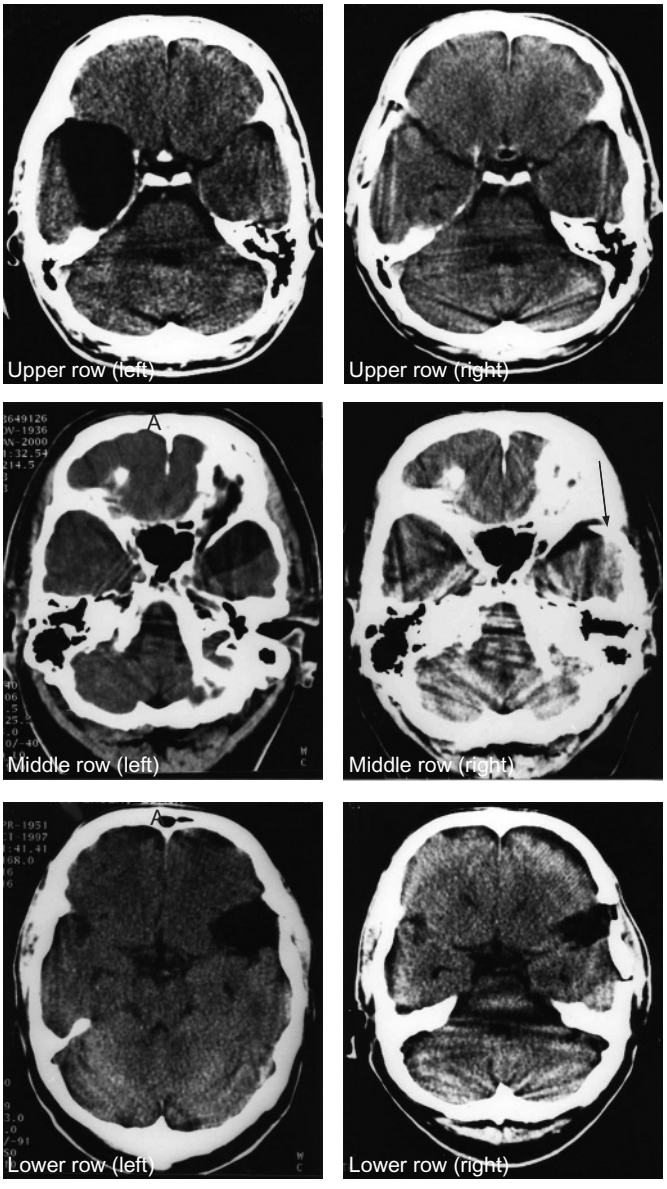

Figure 1 Examples of neuroimaging outcome groups (NOGs) 1–3 after decompression of the temporal arachnoid cysts. Left: Preoperative scan; right: 3–6 months postoperatively. Upper row: NOG1. The cyst is no longer visible on the postoperative control (right). Middle row: NOG2. The postoperative CT scan demonstrates a fluid volume that is less than 50% of the preoperative cyst volume. Note the tip of the intracystic catheter on the postoperative image (arrow). Lower row: NOG3. The postoperative volume is clearly reduced but the residual volume is above 50% of the preoperative cyst volume.

In a few cases, exact neuroimaging categorisation was difficult to decide. The postoperative neuroimaging result was then always assigned to the less favourable group.

Response to questionnaires

A total of 153 letters with questionnaires were sent out. Three patients had died from unrelated causes during the follow-up period and were therefore not contacted. Most patients answered after the first letter. Patients who did not answer within a month received a reminder with the questionnaire enclosed. When letters were returned because of an unknown address, we tried to reach the patients by contacting family members. When this also failed, the hospital's address register was regularly checked for a new address until 6 months after the initial letters were sent out. Three patients could not be located and one elderly patient had developed severe dementia (the letter was returned with an explanation note from the spouse). Thus seven patients were not included in the clinical analyses. In total, 149 (97.4%) patients responded to and completed the questionnaire.

Statistical analysis

The statistical analyses were performed with SPSS 13.0 for Windows. An α-level of 0.05 was used for all statistical calculations. Contingency tables were analysed with Fisher's exact test or χ^2 statistics.

RESULTS

Postoperatively, all patients showed either clinical symptom relief, a reduction in cyst volume, or both. Thus a satisfactory clinical and/or neuroimaging result was obtained in all patients. For all cyst locations, clinical improvement was observed for all cyst sizes, even the smallest. Thus there was no cyst size below which surgical decompression did not seem helpful.

Temporal cysts

Of the 108 patients (96%) with a temporal cyst that completed the questionnaire, 54 (50.0%) were symptom-free at follow-up (COG1). Thirty-three (30.6%) patients reported a significant reduction in their preoperative complaint(s) (COG2) whereas 13 (12.0%) reported no change after operation (COG3).

Eight patients (7.4%) experienced postoperative worsening of their symptoms (COG4). Their characteristics are summarised in table 3.

Headache as the presenting complaint predicted a favourable outcome (COG1 or COG2) ($p = 0.002$). No other symptoms, cyst type (Galassi), method of operation (shunt or fenestration) or occurrence of complications/treatment failure was significantly associated with a favourable/unfavourable clinical outcome in patients with temporal cysts.

Either the preoperative or postoperative imaging could not be found in 14 patients, and therefore these are not included in the neuroimaging analysis. In the remaining 99 patients with a temporal cyst, the cyst was no longer visible on the post-operative images (NOG1) in 28 patients (28.3%) or was reduced to less than 50% of the preoperative volume (NOG2) in 32 (32.3%). Another 26 patients (26.3%) also had a postoperative volume reduction but with a residual volume larger than 50% of the preoperative volume (NOG3). Thus in 13 patients (13.1%) no change in cyst size was observed after operation (NOG4). For the temporal cysts, there were significantly more patients

Table 3 Characteristics of patients with a poor clinical outcome (clinical outcome group 4)

Patient No	Complaint	Cyst location (side)	Galassi type§	Neuroimaging Outcome Group
1*	Headache	Temporal (R)	I	1. Cyst disappeared
2	Headache/dizziness	Temporal (R)	I	2. <50%
3	Headache	Temporal (L)	I	1. Cyst disappeared
4	Headache	Temporal (L)	I	3. >50%
5†	Dementia	Temporal (L)	I	2. <50%
6	Visual disturbance	Temporal (R)	I	2. <50%
7*	Psychiatric disorder	Temporal (R)	II	3. >50%
8‡	Headache	Temporal (L)	III	
9‡	Headache	Posterior fossa (R)		

*Postoperative hygroma, operated on in patient No 7, spontaneously resolved in patient No 1.
†Postoperative subdural empyema.
‡Postoperative neuroimaging examination missing.
§See Galassi and colleagues.[19] Galassi type I is a small cyst confined to the anterior portion of the middle fossa. Type II is larger and has its superior extension along the Sylvian fissure, with displacement of the temporal lobe and the opercula, opening the fissure in a square-like fashion and exposing the insula. Type III fills the entire middle cranial fossa and extends above the middle fossa, displacing the frontal, temporal and parietal lobes.

with an unchanged postoperative volume than for other cyst locations (frontal, posterior fossa and other, different locations) ($p = 0.02$). We could not identify any factor (eg, cyst type, operation method, occurrence of complications) that predicted the final neuroimaging outcome.

Frontal cysts

Of the 22 patients that answered the questionnaire (91.7%), 12 (54.5%) were symptom-free (COG1) after the operation. Nine patients (40.9%) had experienced a significant improvement (COG2) while one patient had no change after surgery (COG3). No patient with a frontal cyst experienced clinical worsening after operation. No factors were found that could predict the clinical outcome after surgery.

In three patients with a frontal cyst, either preoperative or postoperative imaging could not be found. In the remaining 21 patients (87.5%), the cyst had disappeared (NOG1) in 13 (61.9%) or was reduced to less than 50% (NOG2) in the remaining eight.

Volume reduction to less than 50% of the preoperative volume (NOG1 and 2) was significantly more frequent in patients with frontal cysts than in patients with all other cyst locations ($p = 0.002$). No factors were identified that predicted the postoperative volume reduction for patients with frontal cysts.

Posterior fossa

All patients with cysts in the posterior fossa completed the questionnaire. Five patients were symptom-free (COG1) while five had a significant reduction in their preoperative complaints (COG2). Two patients had no change in their symptoms (COG3) while one patient (table 3) experienced postoperative worsening (COG4). Dizziness/nausea as the presenting complaint was associated with a favourable outcome (COG1 and/or COG2) ($p = 0.045$).

Imaging was unavailable in one patient. In the 11 patients eligible for radiological analysis, the cyst was no longer visible (NOG1) in one. In three patients, the cyst was reduced to less than 50% (NOG2), while in seven patients it was reduced but the reduction was less than 50% (NOG3). Volume reduction to less than 50% of preoperative volume (NOG1 and 2) was less frequent for patients with cysts in the posterior fossa than for patients with other cyst locations ($p = 0.035$).

Other locations

Six patients had cysts in a location other than the temporal, frontal or posterior fossa, but they were too few and too heterogeneous to be analysed separately.

Association between clinical and neuroimaging results

For the 133 patients where both the clinical and neuroimaging outcome could be assessed, a good clinical outcome (COG1 and 2) was associated with a good neuroimaging outcome (NOG1 and 2) in 75 patients (56.4%), and with a less favourable neuroimaging outcome (NOG3 and 4) in 37 (27.8%). However, 14 of the patients with a less favourable outcome (COG3 and 4) had a good neuroimaging outcome (NOG1 and 2). Only seven patients with a poor clinical outcome (COG3 and 4) also had a poor neuroimaging outcome (NOG3 and 4). Thus there was no difference in clinical outcome between the patients who had a postoperative volume <50% and those where the postoperative volume was >50% of the preoperative cyst volume. This lack of association between neuroimaging and clinical results was observed for all cyst locations and for all major symptoms.

Self-reported level of function

A total of 97 patients (66%) reportedly functioned better after surgery. Forty (27%) reported no change and 11 patients (7%) reported worsening of overall function after operation. One respondent did not complete that part of the questionnaire.

Of the 11 patients who reported worsening of overall function after surgery, two were in COG2, one in COG3 and eight in COG4. The symptoms of these patients were headache in six, epilepsy in one, psychiatric disorder in one, visual disturbance in two and dementia in one. Preoperative or postoperative imaging was missing in two of these patients. In the remaining nine patients, the cyst was no longer visible (NOG1) in three, reduced to <50% (NOG2) in five, and reduced, but still >50% of the preoperative volume (NOG3), in one. Thus all 9 patients with worsening, where it also was possible to compare preoperative and postoperative volumes, had some degree of volume reduction.

Complications and failures

Failures were defined as lack of effect of treatment whereas complications were defined as additional pathological conditions caused by the treatment. None of the complications encountered were severe. A complication caused a minor, permanent invalidity in only two patients, a persistent trochlear nerve paralysis in a patient with a type II temporal cyst and a postoperative anosmia in one patient, operated on for a type III cyst

Complications

Complications occurred in 26 patients (16.9%), necessitating additional surgery in 11 patients, burr holes under local anaesthesia in the majority (table 4).

Table 4 Complications

Complication	No of patients	No reoperated	No of procedures
Subdural haematoma*	13	7	14
Subdural hygroma	8	2	3
Subdural empyema	1	1	1
Epidural haematoma†	1	1	1
IV cranial nerve palsy‡	3	1†	1†
Anosmia	1		

*External hydrocephalus in one patient.
†This patient had a IV cranial nerve palsy and an epidural haematoma.
‡Temporary in two, permanent in one.

Table 5 Reoperation because of treatment failure

Reoperation method	Primary operation	
	Craniotomy	Internal shunt
Craniotomy	6	6
Revision of shunt		2
New internal shunt	2	4
Sum of interventions	8*	12†

*One patient was reoperated initially with craniotomy, and subsequently with two insertions of an internal shunt.
†One patient was reoperated twice with insertions of an internal shunt, followed by a craniotomy. One patient was reoperated with revision of the shunt and subsequently with insertion of a new internal shunt.

Patients with a temporal cyst seemed especially prone to complications, as all patients with a complication, regardless of complication type, had a temporal cyst ($p<0.001$). Cyst size did not influence the complication rate, as patients with small and medium sized cysts (types I and II) were equally disposed to complications as patients with type III cysts. There was no difference in complication rate between the surgical methods (craniotomy with fenestration vs insertion of an internal shunt; Fisher $p = 0.299$). However, for the most common complication, postoperative subdural haematoma (SDH), there was a significant difference between the two surgical methods for patients with a temporal cyst. An SDH occurred more frequently following insertion of an internal shunt (5 of 21 patients) than after a craniotomy with fenestration (8 of 92 patients; $p = 0.05$).

In patients who developed a IV cranial nerve palsy, two had spontaneous remission after a few months while it remained permanent in one patient. The anosmia (table 4) occurred in a patient with a large type III cyst, and was probably caused by traction on the olfactory bulbs due to a postoperative brain shift towards the cyst side. In a few patients, we observed a moderate, partial facial nerve palsy affecting the ipsilateral forehead. This caused no major problems for the patients, and was not seen more frequently than after other craniotomies.

Failures

Fifteen patients (10 males, 5 females) were reoperated because of suspected or established treatment failure. Patients that were primarily operated on with the minimally invasive internal shunt procedure were more prone to treatment failure than those operated on with a craniotomy with fenestration (9 of 34 with internal shunt vs 6 of 120 with a fenestration through a craniotomy; Fisher $p = 0.001$). The failure rate was not associated with cyst location. Six patients that were initially operated on with a craniotomy had a second craniotomy performed. One of these patients subsequently had two internal shunts inserted. Nine patients who initially were operated on with insertion of an internal shunt were also reoperated. In five of these patients, the second procedure was a craniotomy. One patient was reoperated twice with insertion of a new internal shunt, and finally with a craniotomy. Two patients had a revision of their internal shunts. In one, a new shunt was later inserted. One patient was reoperated once with insertion of a new internal shunt. The reoperations are summarised in table 5.

DISCUSSION

Overall, the majority of patients operated on for an arachnoid cyst had a good clinical and neuroimaging outcome. In addition, most patients reported better overall function after surgery. This was accomplished without any severe permanent complications related to the treatment.

In separate studies, including subgroups of these patients, we have previously demonstrated dyscognition in patients with frontal and temporal cysts, and that these cognitive deficits disappear after surgical cyst decompression.[6 9 10] A similar dyscognition and postoperative normalisation have also been documented in other studies.[3 5 8 21] Arachnoid cysts have also been demonstrated to influence metabolism and perfusion of the adjacent cerebral tissue, with normalisation after surgery.[4 7]

Thus we believe that the clinical improvement seen in most of our patients combined with the documented dyscognition that is reversed by surgical cyst decompression indicates that the previously rather common attitude of reluctance to operate on such patients perhaps should be questioned, and that a more liberal indication for surgical decompression of arachnoid cysts should be adopted. However, intracranial surgery with the aim of improving the quality of life should only be undertaken if the surgical risk is low. Our results indicate that it is.

It is an interesting finding that neuroimaging improvement did not parallel, nor did it seem to be a prerequisite for, clinical improvement, or vice versa: we had some patients with a clear neuroimaging improvement, but with persisting complaints, and we observed clinical improvement in others without a corresponding neuroimaging change. This is in contrast with our findings in children where we found a clear correlation between neuroimaging and clinical results.[17] A similar dissociation between clinical improvement and neuroimaging results has been reported by some authors[8 22] whereas others have found that neuroimaging and clinical improvement are closely linked.[8 9 11 13]

This lack of association between clinical and neuroimaging outcomes may be explained by the following. The neurocranium is moulded after its content. Thus the presence of a congenital arachnoid cyst during the growth of the skull will create a surplus intracranial space, such as the commonly observed enlarged middle fossa in patients with temporal fossa cysts. Consequently, there will be a mismatch between skull volume and brain volume, and the resulting intracranial volume may be too large for the relieved brain to fill completely, even after successful cyst decompression, because the brain simply is not large enough.[23–26] In children, this mismatch between skull and brain volumes may not be as pronounced, provided they are operated on while the brain (and the skull) is still growing.

Another interesting finding is that in the group of patients with the worst clinical outcome (COG4), most patients were operated on for a small cyst and had a good neuroimaging outcome. One possible explanation may be that their complaints were unrelated to their cyst, an explanation that seems probable in retrospect. Headache is the most common complaint in cyst patients, but it is also a symptom that can have many other causes. We found that most of our patients had good clinical relief, with a low risk of complications after surgical decompression. We therefore decided to operate without subjecting them to a series of preoperative investigations, to find other possible causes of headache. Some of the

COG4 patients may thus have had other causes for their headache.

A number of different treatment modalities for arachnoid cysts exist, and the advantages and disadvantages of the individual methods have been thoroughly discussed.[20 22 25 27–31] The majority of our patients were operated on with a fenestration of the cyst to the basal cisterns through a craniotomy. This gave a favourable outcome in most of the patients, with complication rates similar to other reports in the literature.[25 31–36] Fenestrating the cyst wall by an open craniotomy or by endoscopic procedures probably equals out the pressure difference between the cyst and the rest of the intracranial compartment, as also is the case with insertion of an internal shunt.[22 30] It is reasonable to believe, on the other hand, that shunting the cyst content to the peritoneum in most cases will create a new intracranial pressure gradient, as the intracystic pressure is selectively lowered compared with the rest of the intracranial compartment. From this point of view, we therefore believe that fenestration or the internal shunt techniques are to be preferred in the treatment of intracranial arachnoid cysts.

In our material, complications occurred only in cysts located in the temporal fossa, and the most common complication, postoperative SDH, occurred more frequently in patients treated with a shunt than in patients who underwent a craniotomy. From the literature it is well known that temporal cysts predispose to chronic SDHs.[15 37] The occurrence of chronic SDH after insertion of an internal shunt can be because of shifting of brain structures and traction on the bridging veins, thus inducing leakage of blood from the point of the vein's entry into the basal sinuses, an observation we often have made during our open cyst operations.

We have made two additional observations that may be of interest in this context. Firstly, we have regularly noticed how easily the parietal arachnoid cyst membrane is detached from the dura in the middle fossa. Just lowering the intracystic pressure by opening the cyst may cause the membrane to collapse away from the dura. Secondly, when this occurs, the unveiled dura often begins to ooze blood from multiple small openings in the dural surface. The loosely attached parietal membrane thus seems to serve as an extra wall of vascular structures in the dura. Any treatment modality that lowers intracystic pressure without controlling for this dural surface bleeding may therefore be more likely to cause a postoperative SDH. This is probably the reason for our higher occurrence of SDH in shunt patients than in craniotomy patients, where the dural bleeding could be observed, and haemostasis could be achieved. Endoscopic procedures also cause the cyst to collapse much in the same way as a shunt procedure does. However, it is not known whether such procedures are associated with an increased occurrence of postoperative SDH.

We have also operated on a limited number of patients using the internal shunt technique. Although this technique is a relatively simple, safe and efficient method, we tend to reserve this as an alternative method of treating patients with arachnoid cysts where a full craniotomy under general anaesthesia is not recommended. This reservation is because of the observed higher failure rate of this method compared with fenestration.

CONCLUSION

Treatment of patients with arachnoid cysts, including those presenting with unspecific symptoms, carries a clear clinical benefit for patients, and can be performed with little risk of severe complications. These results, based on a rather liberal indication for surgery, indicate that an increased number of patients with intracranial arachnoid cyst would benefit from surgical cyst decompression.

Authors' affiliations

Christian A Helland, Knut Wester, Section for Neurosurgery, Department of Surgical Sciences, University of Bergen, and Department of Neurosurgery, Haukeland University Hospital, Bergen, Norway

Competing interests: None.

REFERENCES

1 **Wester K**. Peculiarities of intracranial arachnoid cysts: Location, sidedness, and sex distribution in 126 consecutive patients. *Neurosurgery* 1999;**45**:775–9.
2 **Clavel M**, Taborga FG, Onzain I. Arachnoid cysts as a cause of dementia in the elderly. *Acta Neurochir (Wien)* 1985;**78**:28–32.
3 **Colameco S**, DiTomasso RA. Arachnoid cyst associated with psychological disturbance. *Journal Med Soc N J* 1982;**79**:209–10.
4 **De Volder AG**, Michel C, Thauvoy C, *et al*. Brain glucose utilisation in acquired childhood aphasia associated with a sylvian arachnoid cyst: recovery after shunting as demonstrated by PET. *J Neurol Neurosurg Psychiatry* 1994;**57**:296–300.
5 **Lang W**, Lang M, Kornhuber A, *et al*. Neuropsychological and neuroendocrinological disturbances associated with extracerebral cysts of the anterior and middle cranial fossa. *Eur Arch Psychiatry Neurol Sci* 1985;**235**:38–41.
6 **Raeder MB**, Helland CA, Hugdahl K, *et al*. Arachnoid cysts cause cognitive deficits that improve after surgery. *Neurology* 2005;**64**:160–2.
7 **Sgouros S**, Chapman S. Congenital middle fossa arachnoid cysts may cause global brain ischaemia: a study with 99Tc-hexamethylpropyleneamineoxime single photon emission computerised tomography scans. *Pediatr Neurosurg* 2001;**35**:188–94.
8 **Soukup VM**, Patterson J, Trier TT, *et al*. Cognitive improvement despite minimal arachnoid cyst decompression. *Brain Dev* 1998;**20**:589–93.
9 **Wester K**, Hugdahl K. Arachnoid cysts of the left temporal fossa—impaired preoperative cognition and postoperative improvement. *J Neurol Neurosurg Psychiatry* 1995;**59**:293–8.
10 **Wester K**, Hugdahl K. Verbal laterality and handedness in patients with intracranial arachnoid cysts. *J Neurol* 2003;**250**:36–41.
11 **Erman T**, Göçer AI, Tuna M, *et al*. Intracranial arachnoid cysts. Clinical features and management of 35 cases and review of the literature. *Neurosurg Q* 2004;**14**:84–9.
12 **Koch CA**, Voth D, Kraemer G, *et al*. Arachnoid cysts: does surgery improve epileptic seizures and headaches? *Neurosurg Rev* 1995;**18**:173–81.
13 **Mayr U**, Aichner F, Bauer G, *et al*. Supratentorial extracerebral cysts of the middle cranial fossa. A report of 23 consecutive cases of the so-called temporal lobe agenesis syndrome. *Neurochirurgia (Stuttg)* 1982;**25**:51–6.
14 **Sommer IE**, Smit LM. Congenital supratentorial arachnoidal and giant cysts in children: a clinical study with arguments for a conservative approach. *Childs Nerv Syst* 1997;**13**:8–12.
15 **Parsch CS**, Krauss J, Hofmann E, *et al*. Arachnoid cysts associated with subdural hematomas and hygromas: analysis of 16 cases, long-term follow-up, and review of the literature. *Neurosurgery* 1997;**40**:483–90.
16 **van der Meche FG**, Braakman R. Arachnoid cysts in the middle cranial fossa: cause and treatment of progressive and non-progressive symptoms. *J Neurol Neurosurg Psychiatry* 1983;**46**:1102–7.
17 **Helland CA**, Wester K. A population-based study of intracranial arachnoid cysts. Clinical and radiological outcome following surgical cyst decompression in children. *J Neurosurg* 2006;**105**:385–90.
18 **Helland CA**, Wester K. Arachnoid cysts in adults: long-term follow-up of patients treated with internal shunts to the subdural compartment. *Surg Neurol* 2006;**66**:56–61.
19 **Galassi E**, Tognetti F, Gaist G, *et al*. CT scan and metrizamide CT cisternography in arachnoid cysts of the middle cranial fossa: classification and pathophysiological aspects. *Surg Neurol* 1982;**17**:363–9.
20 **Wester K**. Arachnoid cysts in adults: Experience with internal shunts to the subdural compartment. *Surg Neurol* 1996;**45**:15–23.
21 **Zaatreh MM**, Bates ER, Hooper SR, *et al*. Morphometric and neuropsychologic studies in children with arachnoid cysts. *Pediatr Neurol* 2002;**26**:134–8.
22 **McBride LA**, Winston KR, Freeman JE. Cystoventricular shunting of intracranial arachnoid cysts. *Pediatr Neurosurg* 2003;**39**:323–9.
23 **Garcia-Bach M**, Isamat F, Vila F. Intracranial arachnoid cysts in adults. *Acta Neurochir Suppl (Wien)* 1988;**42**:205–9.
24 **Geissinger JD**, Kohler WC, Robinson BW, *et al*. Arachnoid cysts of the middle cranial fossa: surgical considerations. *Surg Neurol* 1978;**10**:27–33.
25 **Kandenwein JA**, Richter HP, Borm W. Surgical therapy of symptomatic arachnoid cysts—an outcome analysis. *Acta Neurochir (Wien)* 2004;**146**:1317–22.
26 **Koch CA**, Moore JL, Voth D. Arachnoid cysts: how do postsurgical cyst size and seizure outcome correlate? *Neurosurg Rev* 1998;**21**:14–22.
27 **Arai H**, Sato K, Wachi A, *et al*. Arachnoid cysts of the middle cranial fossa: experience with 77 patients who were treated with cystoperitoneal shunting. *Neurosurgery* 1996;**39**:1108–12.
28 **Choi JU**, Kim DS, Huh R. Endoscopic approach to arachnoid cyst. *Childs Nerv Syst* 1999;**15**:285–91.

29 **Levy ML**, Wang M, Aryan HE, *et al*. Microsurgical keyhole approach for middle fossa arachnoid cyst fenestration. *Neurosurgery* 2003;**53**:1138–44.
30 **Sato H**, Sato N, Katayama S, *et al*. Effective shunt-independent treatment for primary middle fossa arachnoid cyst. *Childs Nerv Syst* 1991;**7**:375–81.
31 **Samii M**, Carvalho GA, Schuhmann MU, *et al*. Arachnoid cysts of the posterior fossa. *Surg Neurol* 1999;**51**:376–82.
32 **Kang JK**, Lee KS, Lee IW, *et al*. Shunt-independent surgical treatment of middle cranial fossa arachnoid cysts in children. *Childs Nerv Syst* 2000;**16**:111–16.
33 **Lena G**, Erdincler P, Van CF, *et al*. Arachnoid cysts of the middle cranial fossa in children. A review of 75 cases, 47 of which have been operated in a comparative study between membranectomy with opening of cisterns and cystoperitoneal shunt. *Neurochirurgie* 1996;**42**:29–34.
34 **Nowoslawska E**, Polis L, Kaniewska D, *et al*. A comparison of neuro-endoscopic techniques with other surgical procedures in the treatment of arachnoid cysts in children. *Neurol Neurochir Pol* 2003;**37**:587–600.
35 **Raffel C**, Mccomb JG. To shunt or to fenestrate: which is the best surgical treatment for arachnoid cysts in pediatric patients? *Neurosurgery* 1988;**23**:338–42.
36 **Tamburrini G**, Caldarelli M, Massimi L, *et al*. Subdural hygroma: an unwanted result of Sylvian arachnoid cyst marsupialization. *Childs Nerv Syst* 2003;**19**:159–65.
37 **Page AC**, Mohan D, Paxton RM. Arachnoid cysts of the middle fossa predispose to subdural haematoma formation fact or fiction? *Acta Neurochir Suppl (Wien)* 1988;**42**:210–15.

HISTORICAL NOTE

doi: 10.1136/jnnp.2007.119669

Pierre Mollaret (1898–1987) and his legacy to science

Pierre Mollaret was a French physician who made various significant contributions to neurology and infectious diseases. He was born in Auxerre, France in 1898. In 1916, he began to study medicine and science but his education was interrupted by World War I. During 1917 and 1918, he served as an assistant physician and was decorated with the Croix de Guerre when the war ended. He resumed his medical studies in 1920 and received his degree in science in 1926. During his education, one of his teachers was Professor Georges Charles Guillian (1876–1961; Guillian of the Guillian–Barré syndrome), with whom Mollaret worked for many years.

In 1931, Guillain and Mollaret described anatomical connections related to palatal myoclonus.[1] This functional connection, composed of the contralateral dentate nucleus, ipsilateral red nucleus and ipsilateral inferior olivary nucleus, is referred to as the Guillain–Mollaret triangle.[2] In fact, it is not an anatomical triangle but a triangular circuit connecting the aforementioned structures. In 1935, Trelles (1904–1990), a Peruvian neurologist, reported that isolated lesions of the inferior cerebellar peduncle never cause palatal myoclonus, as anatomically there are no direct connections between the inferior olivary nucleus and the contralateral dentate nucleus. Fibres from the inferior olivary nucleus project first to the cerebellar cortex (olivocerebellar tracts) and then to the dentate nucleus. He described these connections as the dentato-rubral-olivary pathway.[3]

In the central nervous system, degeneration of an anatomical structure is usually characterised by neuronal loss replaced by proliferation of glial elements. Degeneration of the inferior olivary nucleus was described for the first time in 1887 by Hermann Oppenheim (1858–1919) from anatomical specimens.[4] Hypertrophic olivary degeneration is considered a unique type of degeneration because it is associated with enlargement of the affected structure. Hypertrophic olivary degeneration is a rare finding secondary to focal lesions, usually an insult, of the brainstem involving a functional system and is considered a transneuronal degeneration. Transneuronal degeneration occurs only from a lesion that results in disconnection of the pathway and loss of neuronal input. While hypertrophic olivary degeneration can be caused by any lesion involving the aforementioned structures, it is typically seen with focal lesions involving the ipsilateral central tegmental tract, the contralateral superior cerebellar peduncle or the dentate nucleus.

In 1944, Mollaret also described three patients with recurrent benign endothelioleukocytic aseptic meningitis.[5] He wrote several subsequent papers on the condition. The European literature added additional cases, but reports of this disease did not appear in the English language medical literature until 1972.[6] It has also been called benign recurrent aseptic meningitis, benign recurrent meningitis, benign recurrent endothelial meningitis, benign recurrent endothelial–leukocytic meningitis and recurrent aseptic meningitis.[7]

Mollaret was also an epidemiologist. He held the chair of Infectious Diseases at the Claude Bernard Hospital in Paris and contributed towards the care of a large number of polio victims suffering from respiratory paralysis.[8] Mollaret as a neurologist and epidemiologist made innovative contributions to science. He died in Montgeron, near Paris, in 1987, and left behind many articles and books.

Levent Sarikcioglu, Muzaffer Sindel
Department of Anatomy, Akdeniz University Faculty of Medicine, Antalya, Turkey

Correspondence to: Dr Levent Sarikcioglu, Department of Anatomy, Akdeniz University Faculty of Medicine, 07070 Antalya, Turkey; sarikcioglu@akdeniz.edu.tr

Competing interests: None.

References

1 **Guillain G**, Mollaret P. Deux cas de myoclonies synchrones et rythmées vélo-pliaryngo-oculo-diaphragmatiques. Le problème anatomique et physio-pathologique de ce syndrome. *Rev Neurol (Paris)* 1931;**2**:545–66.
2 **Gerace C**, Fele MR, Luna R, *et al*. Neurological picture. Bilateral hypertrophic olivary degeneration. *J Neurol Neurosurg Psychiatry* 2006;**77**:73.
3 **Salamon-Murayama N**, Russell EJ, Rabin BM. Diagnosis please. Case 17: hypertrophic olivary degeneration secondary to pontine hemorrhage. *Radiology* 1999;**213**:814–17.
4 **Venturi F**, Tampieri D, Brassard R, *et al*. Hypertrophic olivary degeneration (HOD): an unusual way to degenerate. *Neuroimage* 2001;**17**:7–8.
5 **Mollaret P**. La méningite endothélio-leucocytaire multirécurrente benigne: syndrome nouveau ou maladie nouvelle? *Rev Neurol (Paris)* 1944;**76**:57–67.
6 **Ruben SJ**. Mollaret's meningitis. *West J Med* 1994;**160**:459–62.
7 **Shalabi M**, Whitley RJ. Recurrent benign lymphocytic meningitis. *Clin Infect Dis* 2006;**43**:1194–7.
8 **Seytre B**. *Death of a disease: A history of the eradication of poliomyelitis*. New Brunswick, New Jersey: Rutgers University Press, 2005.